Syphilis and Blindness

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POINTS out the urgency of more interest in syphilis as a systemic disease, in its public health aspects, its treatment, and in early diagnosis of optic nerve treatment.

STATISTICAL studies of the causes of blindness attribute 10 to 15 per cent to syphilis.[1] Hurlin[2] estimated the rate of blindness per 1,000 of population in the United States as 1.7. He concluded that about 230,000 was a reasonable estimate of total number of blind persons in the United States in 1940. On the basis of these figures it can be deduced that there are 23,000 to 34,500 syphilitic blind in the United States.

The outstanding causes of blindness of syphilitic origin are primary atrophy of the optic nerve of acquired and of congenital syphilis and interstitial keratitis of congenital syphilis. Other less common ocular syphilitic lesions that may result in blindness were discussed elsewhere[3] and will not now be considered.

In 1932 Moore[4] estimated that there must be 50,000 cases of primary atrophy of the optic nerve (of acquired syphilis) in the United States. This figure was based on an estimate of 423,000 new cases of syphilis each year, the census figures, and the estimate that 10 per cent of the adult population is infected with syphilis. Of untreated or poorly treated syphilitics, approximately 5 per cent develop tabes. There are about one half million persons with tabes in the United States and 10 to 15 per cent of these may be expected to develop optic nerve atrophy.

Since Moore's estimate the census has shown an increase. On the other hand the incidence of new cases of syphilis has decreased. The estimate

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for 1947 in the United States is 250,000.[5] According to Dr. Walter J. Clarke, American Social Hygiene Association (personal communication), the estimated annual incidence of new cases, which prior to 1936 was 500,000, had been reduced to approximately 220,000 by 1945. Since the end of the war the incidence appears to have risen to about 250,000 cases each year.

Primary Syphilitic Atrophy of the Optic Nerve

Acquired Syphilis

Syphilitic primary* atrophy of the optic nerve of acquired syphilis is associated with neurosyphilis. It therefore occurs with the different types of neurosyphilis, especially the tabetic type.

A study[3] of 377 patients with optic nerve involvement revealed that of this number 20 had congenital syphilis; in the remainder the infection was acquired. Males, both white and Negro, were more affected than females, in a ratio of 3.5 to 1. The youngest was 24, the oldest was 80. The predominating decade when first seen (for reason of impaired vision) was 40 to 50.

An unfortunate observation in our study, which we wish to stress, is that 61 per cent of our 377 patients with acquired syphilis when first seen were either industrially blind, 6/60 (20/200) in the better eye (the upper limit for industrial blindness), or had visual acuity of less than 1/60 (3/200). Only 11.3 per cent had visual acuity of 6/9–6/6 (20/30–20/20); and in only 13.3 per cent it was 6/12–6/15 (20/40–20/50).

In Lehrfeld and Gross's[6] study of optic atrophy of patients attending Wills Eye Hospital in Philadelphia, it was determined that of 338 patients there was a history of a chancre in 26 per cent and in 14 per cent a history of having had gonorrhea. The remaining 60 per cent had no knowledge of infection. The patients in this series received either grossly inadequate or no treatment in the early stage of their infection. Of patients who had knowledge of infection there was an average interval of 18 years between infection and onset of loss of vision, and an average of 18 months between onset of loss of vision and institution of what might be regarded as proper treatment of optic atrophy.

An important observation of Lehrfeld and Gross was that 31 per cent of their patients at the onset of impaired visual acuity consulted an optometrist as an initial means of treatment. Some visited the same or a different optometrist for as long a period as two years. It was exceptional that the optometrist referred the patient to the proper place for study. Other patients stated that as an initial step in treatment they consulted an "eye doctor"; when further questioned, however, they said the "eye doctor" had a store and dispensed glasses. The majority of clinic clientele did not know the difference between an optometrist and an oculist or ophthalmologist.

In a considerable percentage of our patients the underlying neurosyphilis pursued a silent course, so far as subjective symptoms were concerned. Frequently impaired visual acuity was

* The word "primary" indicates that the atrophy is the initial process affecting the nerve, whereas in secondary atrophy, some preceding eye disorder induced a resultant atrophy.

the first and only symptom that motivated the patient to seek medical consultation and was therefore the first symptom that led to the diagnosis of the underlying infection.

Atrophy of the optic nerve is rarely associated with other frank subjective symptoms of neurosyphilis, except for pain in the legs typical of the tabetic type of neurosyphilis, although it was usually not incapacitating or sleep-disturbing and was in many patients elicited only upon questioning. Diplopia (double vision) was another accompanying symptom of atrophy of the optic nerve. It was usually transitory and invariably preceded, frequently by years, complaint of visual loss.

Duration of Impaired Vision

The duration of impaired visual acuity of the untreated patients in our series varied considerably from a few weeks to months and years. Rarely had symptoms been present more than five to ten years. At times the patient suddenly became cognizant of impaired vision. Typical of the comments are: "My eyes were good until six months ago—all at once when cutting pipe my boss told me I was cutting it crooked." "While driving a car about six months ago, a film came over my left eye." When first seen, the visual acuity of this patient was hand movements in the right eye and light perception in the left.

The degree of visual loss and advancement of optic atrophy existing before the patient becomes aware of impaired sight may be considerable. It is surprising how much impairment of central vision or constriction of peripheral vision may exist before the patient is compelled to stop work. One patient was still working as a general laborer when he came to the clinic. The visual acuity in the right eye was 2/60, in the left eye 6/60.

Some patients became blind in one year or less from the time of their first complaint of impaired vision. These rapidly progressive cases of optic atrophy contributed to the 17 per cent of patients with visual acuity of less than 1/60 (3/200) on admission.

Treatment—Importance of Early Diagnosis

Results[7] of treatment of optic nerve atrophy show that fever treatment especially (induced malaria or vaccines intravenously), combined with penicillin, was effective in some patients in preventing unfavorable progress of the disease. In others, however, despite considerable treatment blindness ensued. In still others the disease became arrested at considerably reduced levels of visual acuity.

Another study[8] reported on patients with neurosyphilis who had normal visual acuity, normal color of the optic discs but who had, on repeated examinations, perimetric field defects. Following treatment such defects disappeared. This was regarded as the earliest evidence of optic nerve involvement—the preatrophic stage preceding degeneration or atrophy of the optic nerve—the atrophic stage. In the latter stage there is some impairment of visual acuity, pallor of the optic discs, and some degree of perimetric field defects. It is very likely that with careful perimetric studies, the diagnosis of optic nerve involvement could be

made in the pre-atrophic stage before the development of the irrevocable impairment of vision observed in the well-established atrophic stage.

To accomplish this early diagnosis necessitates perimetric field study of every patient with neurosyphilis. Such examination should therefore be a routine one. It will then also serve to diagnose optic nerve involvement early in the atrophic stage, before the patient becomes aware of impaired vision.

Optic Atrophy in Congenital Syphilis

As in acquired infection, atrophy of the optic nerve in congenital syphilis is associated with neurosyphilis. According to Igersheimer,[9] 50 per cent of all patients with the tabetic and the paretic forms of neurosyphilis develop optic nerve atrophy.

Atrophy of the optic nerve of congenital syphilis is fortunately much less common than in acquired syphilis. Moreover, as presently discussed, the incidence of congenital syphilis is decreasing. The disease occurs at an earlier age than in the acquired infection. In our series of twenty patients, females predominated (14 of the 20 patients). The youngest was 9 years of age, the oldest was 34 years. The predominating age range when first seen was 13 to 20.

Unless the course of the disease is checked by treatment, blindness is inevitable. These patients remind us of little Hedwig[10] in Ibsen's *Wild Duck:* "Gay and careless and chirping like a little bird she flutters onward into a life of endless night." The implication in the case of Hedwig is congenital syphilis. The cause of impending blindness could be optic atrophy of neurosyphilis or secondary to retinochoroiditis.

Interstitial Keratitis

Interstitial keratitis is an inflammation of the cornea, of syphilitic causation, affecting almost exclusively patients with congenital or hereditary syphilis. It is the second most frequent cause of considerably impaired vision or blindness caused by syphilis.

Females are more affected than males, in a ratio of about 2.5 to 1. Both eyes are invariably affected either in a short space of time or at varying intervals. In a study[11] of 485 patients the predominating age at onset was between the seventh and thirteenth years for females, and the fifth to the twentieth year for the males. Despite considerable treatment, recurrence of the disease is not uncommon, appearing after a variable period from the first attack. In a patient 65 years of age we have observed recurrence 35 years after the initial attack.

The earlier statistics of the final visual acuity of treated patients with interstitial keratitis ranged up to as high as 20 per cent with visual acuity of less than 6/60 (20/200). With more advanced methods of treatment, especially fever, this percentage has been considerably reduced. In a recent study[12] in which fever and penicillin or chemotherapy were employed the percentage of final visual acuity of between 6/60 (20/200) and 1/60 (3/200) of 97 affected eyes of 59 patients was 8.2; and 2.1 where the vision was less than 1/60 (3/200).

In addition to a more favorable outcome of interstitial keratitis on visual acuity compared with optic nerve atrophy is the decreasing incidence of congenital syphilis. We

have noted a decrease in the incidence of the disease at Wills Eye Hospital. This is consistent with the following statistical data. The syphilitic infant mortality rate per 1,000 live births in continental United States in 1933 was 0.79 (both white and nonwhite), whereas in 1948 it was 0.12. The syphilis death rate per 100,000 population for congenital syphilis in 1939 was 1.0, whereas in 1948 it was 0.3.[13]

This decrease in the incidence of congenital syphilis is to be expected as a result of laws requiring premarital Wassermann test and the now current practice of routine Wassermann test in pregnancy, and treatment with penicillin. In consideration of these circumstances congenital syphilis and interstitial keratitis should become an anachronism.

The Problem of Blindness Caused by Syphilis

As aforementioned interstitial keratitis, as part of congenital syphilis, is showing a decreasing incidence and need not be discussed. The problem of optic nerve atrophy of acquired syphilis is of greater magnitude. In our experience there is no evidence of a decrease in its incidence, and therefore it is of grave concern in the prevention of blindness.

The elimination of acquired syphilitic optic nerve atrophy is obviously concerned with measures that decrease the incidence of syphilis, of its spread, its early diagnosis and proper treatment. Another phase is the early recognition of neurosyphilis and its proper treatment. This necessitates routine spinal fluid examination of every person whose infection is of less than about ten years' duration. In this period neurosyphilis is more likely to be diagnosed by spinal fluid examination than by clinical evidence.

Early Recognition Imperative

The early diagnosis of optic nerve involvement, primarily in the preatrophic stage, secondarily in the atrophic stage, should be emphasized. The disease should be diagnosed before it reaches the symptom level of impaired visual acuity, at which time considerable loss of vision may have ensued which cannot be restored by treatment. Diagnosis in this stage is comparable to the diagnosis of cancer by cachexia. Apparently the optometrist is the first to see one third of the early cases of optic atrophy who later attend ophthalmologic clinics. A corollary of this in the prevention of blindness caused by syphilis is not only the education of the public but of the optometrist to recognize pupillary abnormalities as an important symptom of optic atrophy caused by syphilis and to refer the patient to the proper place for study and treatment.

To facilitate early diagnosis of optic atrophy it is necessary that ophthalmologists and all physicians who examine patients with syphilis have a high degree of awareness of the optic nerve. Not only patients with tabes but all patients with neurosyphilis of acquired and of congenital origin should have examination of the optic nerve. In records of private, ward, and clinic patients too infrequently no mention is made of such examination unless the patient complained of visual loss. The ophthalmologists are admirably situated to exclude optic atrophy of every patient examined with syphilis of some years' duration, at least in patients with pupillary abnormalities.

Ophthalmologic Examination in Syphilis Clinics

In syphilis clinics ophthalmologic examination should be made of every patient with neurosyphilis. If routine examination by an ophthalmologist is not possible, physicians attached to the clinic should learn to recognize abnormal pallor of the optic disc and do visual testing. A visual field with the finger by confrontation may furnish a clue to peripheral loss of vision. Selected cases should be referred for more complete examination.

Since optic atrophy occurs predominantly in persons who have no knowledge of infection, industrial and other physicians concerned with routine serologic tests for syphilis of large numbers of persons can aid in early diagnosis. Ophthalmologic examination should be made on persons whose serologic tests are found positive and who present evidence of neurosyphilis, with particular reference to pupillary abnormalities. In the absence of examination by an ophthalmologist, visual acuity, including vision through a pinhole (to approximate corrected vision) and visual field with the finger by confrontation would be better than no examination.

The 10 to 15 per cent of all blindness caused by syphilis (largely atrophy of the optic nerve) approaches that caused by cataract and glaucoma. Syphilis as a cause is deserving of as much attention as is now being given, for example, to glaucoma and to the publicity in the field of industry of the vital importance of the use of guards and goggles in prevention of injury to the eye.

Public Education Needed

We note absence of a comparable educational or propaganda campaign directed toward early recognition of syphilitic optic nerve atrophy. We therefore appeal to ophthalmologists and to the component divisions of the Committee on Statistics of the Blind for more interest in syphilis as a systemic disease, in its public health aspects, its treatment, and in the early diagnosis of optic nerve involvement.

In this day in which the word "syphilis" is no longer tabooed, it would be helpful in an educational campaign to emphasize that syphilis affects the eye as well as other organs and that complete examination should include the optic nerve. This would be far less daring and inoffensive to the aesthetic sense than the implications in the current match cover appeal to the public of the peril of VD.

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